TABLE I. Characteristics of Patients With WM

Case No.	Age/Sex	IgM (g/L)	L chain	WM cells (%)	viscosity (cp)	ALT (mU/ml)	HCV-RNA	Genotype of HCV
1	34F	65.9	κ	17.0	5.16	78	Positive	III
2	44M	54.1	κ	63.2	4.77	11	Negative	(−)
3	73M	34.9	κ	57.0	n.d.	4	Negative	(−)
4	71M	56.8	λ	48.8	2.65	6	Negative	(−)

L chain, type of light chain, WM cells, atypical lymphoplasmacytoid cells in the bone marrow; viscosity, viscosity of the plasma (normal value = 1.72–2.03); ALT, alanine aminotransferase (normal value = 4–30); n.d., not determined.

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Fanconi Aplastic Anemia Associated With β-Thalassemia Trait

To the Editor: The coexistence of two genetic defects both associated with anemia may cause some clinical and hematological abnormalities, different from those found when they are present separately. In populations with a high incidence of β-thalassemia, such as Turkey, the combination of β-thalassemia mutation and another congenital hematological disorder may occur [1–2]. In Turkey, where the incidence of Fanconi's anemia (FA) also seems high, the coexistence of β-thalassemia trait and Fanconi's anemia in a patient was not surprising [3]. Examination of this child indicated changes in some of her hematological parameters during the severe anemia period, caused by FA and during the remission period which gave us some clues about the counter effects of both abnormalities on these hematological parameters.

Case Report

A 10-year-old girl (S.K.) was referred to our unit for evaluation of severe anemia. She was the third product of a consanguineous marriage, and one of her siblings had died previously of anemia and bleeding at age 8. Her past history revealed that she was diagnosed as having patent ductus arteriosis, which was corrected at the age of 15 months. Physical examination at this hospital at the age of 10 years revealed growth retardation and mild microcephaly. Her height was 127 cm, weight 23 kg (both measurements were less that the 3rd percentile for her age), and head circumference 50 cm. She had two café-au-lait spots, and her right thumb was dislocated. The results of laboratory examination of the patient and of her parents are shown in Table I. Karyotype analysis revealed 46 chromosomes with an XX pattern; an increased rate of spontaneous, and induced chromosomal breakage by diepoxy butane (DEB) was observed. The diagnosis of FA was made, and the patient was treated with oxymethalone 2 mg/kg and prednisolone 5 mg/day. The patient responded to this therapy well in 1 year (Table I). DNA analysis of the β-gene revealed heterozygosity for the IVS1-5 G-C mutation. During the 4-year of follow-up period, the patient remained well. Oxymethalon was tapered to 0.5 mg/kg. Some of the hematological values of the patient during the follow-up period are given in Table I.

The patient presented has FA and β-thalassemia trait associated with the IVSI-5 mutation. Absence of microcytosis during the severe anemia episode caused by FA indicated that the majority of the red blood cells produced by precursor cells are of a fetal line in which Hb synthesis was probably unimpaired because of the active synthesis of the γ-chain [4]. This observation supports the previous hypothesis that fetal red cell precursors are the most resistant cell lines of the marrow precursor cells to abnormalities causing bone marrow aplasia [5]. The presence of microcytosis in remission of FA indicated that the effect of thalassemia on red cell volume overcomes the effects of FA on the same parameter. This observation conflicted with the previous knowledge that in FA during remission or before the anemic episode macrocytosis would be present. This assumption may only be valid in FA patients without a coexistent thalassemic determinant.

This study indicates the importance of detailed hematological evaluation in FA, not only in the anemic period, but during remission as well. Such a detailed study will not only help detect the presence of another genetically transmitted hematological abnormality, but will also aid in understanding the countereffects of different genetic disorders when present in a patient.

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TABLE I. Some Laboratory Data of a Patient With Fancon's Anemia and B-Thal Trait and Her Parents

	Age/ Sex	Hb (g/dl)	WBC (mm^3)	MCV (Fl)	Plat.[a] (10^9/L)	Hb A_2[b] (%)	Hb F[c] (%)
Propositus	10F	4.2	3,000	113	20	—	16
	11	12.4	5,300	68	220	3.6	7
	14	11.0	6,000	68	+++	3.7	8
Father	40M	12.0	7,800	56–61	+++	4.0–4.7	0.5–0.6
Mother	38F	12.0	6,800	87	+++	2.6	0.6

[a]+++, sufficient amount with a good clumps.
[b]By microcolumn.
[c]By alkali denaturation.

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Circulating Leukocytes With Ingested Mucin in a Child With Hirschsprung's Disease

To the Editor: Circulating mucin is an uncommon occurrence in the blood, and occurs almost exclusively either in adults with disseminated adenocarcinoma or children with Wilms' tumor [1,2]. It is found extracellularly, resulting in either a falsely high or an erroneously low leukocyte count. Here we report a case with circulating leukocytes showing ingested mucin.

A newborn boy who presented with failure of meconium passage was suspected of having Hirschsprung's disease. On day 5, a barium enema examination was performed, at which time the bowel was accidentally perforated. An emergency laparatomy was performed to repair the perforation. He was put on total parenteral nutrition, and an elective colostomy was performed on day 9. Peripheral blood examination taken shortly after the second operation on day 9 showed a hemoglobin of 14.3 g/dl, platelet count of 176×10^9/L, and leukocyte count of 15.7×10^9/L with 60% neutrophils, 22% lymphocytes, and 18% monocytes. Many of the neutrophils and monocytes showed prominent cytoplasmic vacuoles. Some cells also contained round gray-blue inclusions in their cytoplasm (Fig. 1), which were demonstrated to be mucin by mucicarmine and periodic acid-Schiff (PAS) stains. No cryoglobulin was demonstrated. The patient had an uneventful recovery from the operation, and subsequent peripheral blood examination showed normalization of the leukocyte count and disappearance of the cytoplasmic inclusions. Similar cytoplasmic inclusions could not be found upon review of the peripheral blood smears taken prior to the second operation.

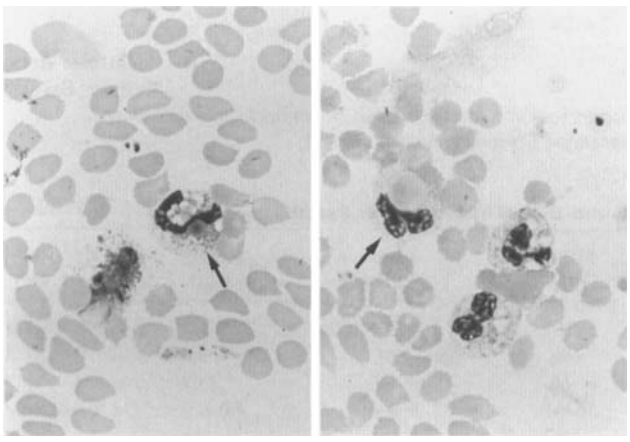

Fig. 1. Composite figure showing neutrophils with ingested mucin (arrow). (Giemsa stain, ×625.)

Cytoplasmic inclusions are sometimes a prominent feature in leukocytes in both reactive and neoplastic conditions. Döhle bodies are collections of rough endoplasmic reticulum characterized by small blue-gray cytoplasmic inclusions that are not uncommonly found in neutrophils in patients who are pregnant or infected [3]. They have, however, a characteristic wedge-shaped appearance and are often situated toward the periphery of the cells. In occasional patients with cryoglobulinemia, neutrophils can contain ingested cryoglobulin [3,4], the appearance of which is often indistinguishable from phagocytosed mucin. However, the cryoglobulin does not react with mucin stains such as mucicarmine and will disappear on warming of the specimen. In view of the clinical history and the temporal relationship with the perforation and repair of the bowel, we speculate that the mucin had gained access to the circulation due to accidental introduction of luminal mucin content or mucin-containing cells into the bloodstream during the operation, with subsequent removal of the mucin by the circulating phagocytes.

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Hemolytic Anemia Precipitated by Pregnancy in a Patient With Hereditary Elliptocytosis

To the Editor: Pregnancy complicated by hemolytic anemia due to hereditary elliptocytosis (HE) occurs extremely rarely [1–4]. Therefore, the maternal and fetal risks in these cases are poorly known. We describe a patient with HE in whom hemolysis was early precipitated and maintained by pregnancy.

A 21-year-old, gravida 1, para 0, white woman presented with weakness, headache, and pallor. She was found as having a 10-week gestation and severe anemia. Her mother and brother were always healthy, but her father, who also was allegedly healthy, was not available for hematologic study. The patient weighed 3,200 g at birth and had shown neonatal jaundice requiring exchange transfusion. Until 3 years of age, she had received blood transfusions, sometimes combined with steroids for anemia several times a year. Afterward she was asymptomatic until her pregnancy.

On physical examination, she had severe pallor and moderate hepatosplenomegaly. The peripheral smear showed many elliptocytes, anisocytosis, and polychromasia. She had reticulocytosis and a raised indirect bilirubin level. Hemoglobin electrophoresis and immunohematologic studies revealed no abnormalities. Following a bone marrow biopsy, hereditary elliptocytosis with hemolytic anemia was diagnosed.

Throughout the prenatal period, the hemoglobin level frequently dropped and jaundice emerged. In addition to iron and folate therapy, the patient